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Title

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Borgerink: acquisition of data and drafting of the manuscript. Visschedijk: critical revision of the manuscript. Weersma: supervision and critical revision of the manuscript.

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Question

A 22-year old woman, born in Pakistan and living in the Netherlands since childhood, presented with progressive bloody diarrhea, severe abdominal pain and vomiting since one month.

Her medical and family history were negative for (gastrointestinal) diseases. Clinical examination revealed an alert woman, without fever or hemodynamic instability. Palpitation of the abdomen was painful without signs of rebound tenderness. Mild hyperpigmentation was shown on the legs and arms. Her laboratory results: hemoglobin 10.9 g/dL, C-reactive protein 44 mg/L, leukocytes 7.1 x 10^9/L, albumin 1.8 g/dL. Liver enzymes and renal values were normal. Fecal and blood cultures remained negative. Alpha-1-antitrypsine was 9,6 mg/g fe, compatible with protein losing enteropathy.

Through colonoscopy we detected multiple polyps seizing almost the complete colonic lumen, causing mild luminal stenosis (Figure 1). A mild inflammation was seen in between the severe polyposis, with a normal mucosal aspect of the ileum and rectum. Biopsies and removal of 1 polyp was performed. Additional abdominal computed tomography revealed diffuse wall thickening of colon and appendix, without signs of perforation or toxic megacolon. Magnetic Resonance Enteroclysis and gastro duodenoscopy detected no abnormalities.

What is most likely underlying in this young adult? What would be the next step in diagnosis and treatment?

Answer to the Clinical Challenges and Images in GI Question:
Inflammatory Bowel Disease, with polyposis and protein-losing enteropathy

Polyposis combined with protein losing enteropathy is most commonly attributed to a polyposis syndrome. In this context, the first differential diagnosis included Cronkhite Canada syndrome (CCS), Peutz Jeghers Syndrome, Juvenile polyposis, Cowden Syndrome, Familiar adenomatous -, MUTH-associated- or CAP-polyposis or Inflammatory Bowel Disease (IBD)(1).

The presentation of this case with bloody diarrhea, protein losing enteropathy, hyperpigmentation and polyposis matched with the CCS(2). Pathology of the first biopsies and polypectomy showed atypical polyps, without hamartomatous or adenomatous features and with mild atypical inflammation. Moreover, a cancer genetic testing panel excluded most of the described causes of polyposis. For these reasons, the differential diagnosis remained CCS and IBD.

Treatment was started with Prednisolone, which failed. The next treatment step was anti-TNF-alpha therapy *(Infliximab)*, which is besides treatment for IBD, also described in case reports for the treatment of CCS(2). It showed minimal improvement and clinical progression was during two doses. Nutritional status was optimized with total parental nutrition. However, the protein-losing enteropathy remained. Despite the treatment, the clinical condition of this patient deteriorated and after previous refusal of a colectomy, she accepted surgical treatment.
Subtotal colectomy with ileostomy was performed three months after presentation, which did reveal crypt distortion of the entire colon (Figure 2). Polyps were histopathologically described as pseudo polyps, with mild inflammation in between and most compatible with ulcerative colitis (Figure 3).
The patient quickly recovered post-surgery. The protein losing enteropathy, abdominal pain and bloody stool disappeared, and she gained weight.

Protein losing enteropathy (PLE) is a rare complication of Inflammatory Bowel Disease, which can only appear after exclusion of liver or kidney failure and malnutrition. Alpha-1-antitrypsine clearance in stool is a rate for PLE(3). In this case, subtotal colectomy was the only treatment left because of clinical decline, but in the end proved to be a solution for PLE.

To conclude, a relatively common disease as Inflammatory Bowel Disease can present with rare severe polyposis with mild inflammation, combined with protein-losing-enteropathy.

References

1. Kanth P, Grimmett J, Champine M, Burt R, Samadder NJ. Hereditary Colorectal Polyposis and Cancer Syndromes: A Primer on Diagnosis and Management. Am J Gastroenterol. 2017;8;112-1509.

2. Watanabe D, Ooi M, Hoshi N, Kohashi M, Yoshie T, Ikehara N, et al. Successful treatment of Cronkhite – Canada syndrome using anti-tumor necrosis factor antibody therapy. Endoscopy. 2014;476–7.

3. Umar SB, DiBaise JK. Protein-Losing Enteropathy: Case Illustrations and Clinical Review. Am J Gastroenterol. 2009;105;43.